

# Butterbean Team



That's our question this year, "Ain't I worth a cure?" For the past several years Team Butterbean has been sending out letters asking for donations to the Cystic Fibrosis Foundation on behalf of our little butterbean, Emily Grace Wilson. Emily, now four, was diagnosed at 5 months with the life shortening genetic disorder Cystic Fibrosis. Last year we reported that Emily received a feeding tube and mediport to aid in her treatments. We also noted she had a set back when she cultured *Pseudomonas*, a nasty lung bug most CFers get at some time in their life. This year we are excited to report that Emily has finally gotten a grasp on her disease. Her weight has exceeded the critical mark of 50% BMI and has stayed there for the entire year. Her normal daily treatments have been tweaked and adjusted such that she has not had a tune-up since December '06 (tune-ups are 2-week courses of IV antibiotics Emily used to receive every three months). Emily has had an excellent year.

But those accomplishments have not come without a lot of hard work. Emily, like most CF kids, spends an hour in the morning and evening doing her breathing treatments which consist of chest percussion therapy and various inhaled medicines. She spends the entire night connected to her feeding tube in order to get the nutritional requirements her little body requires. And then when she starts to get a cold, she requires additional treatments during the day, often given by a loving staff at preschool.

That hard work is a constant reminder that there is no cure, or even direct treatment, for Cystic Fibrosis. While Emily has gotten a grasp for the time being, she still has a hard road to plow. That is why we ask, "Ain't I worth a cure?"

The Cystic Fibrosis Foundation thinks so and raises money to support drug research and development to help treat, and hopefully cure, Cystic Fibrosis. Cystic Fibrosis is an orphan disease with a relative small potential customer base for new drugs. As a result, pharmaceuticals are leery in investing money in risky drug trails with small potential for payout. The Cystic Fibrosis Foundation is not. With every dollar they raise almost 90 cents goes to the cause of finding a cure. An efficiently ratio which ranks the CFF as one of the top charitable research organizations in the nation.

And that research has, and will continue, to pay off in Emily's life. Attached is the Cystic Fibrosis Foundation Therapeutics Pipeline. From this graph you can see there are 5 drugs currently available to CF patients. Emily's daily treatment consists of three of those five, Pulmonzyme, TOBI, and AquaADEK. Hypertonic Saline has been discussed but we have decided to wait till she's older to pursue that treatment. Azithromycin, commonly known as Zithromax, was found to be an effective treatment against *pseudomonas* during a 1999 CFF funded drug trail. Emily takes Azithromycin as the first attack when flair-ups occur. We are excited about the Phase 3 drug Gilead 1020 which should be available in the spring-summer of this year as it may replace TOBI with a quicker, more efficient treatment means.

While those CFF funded drugs have helped Emily immensely this past year, they are what we call reactionary drugs. They are treating the effects of Cystic Fibrosis, not the underlying cause. Because of this, life expectancy can be increased, but the constant drain of living with Cystic Fibrosis is not effected. However, there are multiple drugs in the Phase 3, 2, and 1 stages that will attack this disease head on. Those are the drugs in the first three categories. These are the promising drugs for a cure or suppression of Cystic Fibrosis. This is where you come in.

The research and development of these promising drugs can not be performed with out funding. Please join me in saying "yes, you are worth a cure." by making a donation to the CFF in honor of our little Butterbean, Emily Grace.

Thank you,

Rob and Corinne Wilson

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## Cystic Fibrosis Foundation Therapeutics Pipeline

